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# CONSERVATIVE MANAGEMENT OF IDIOPATHIC CHYLOTHORAX AND CHYLOPERICARDIUM IN A PRETERM NEONATE

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*Chylothorax is a rare condition characterized by accumulation of chyle within the pleural cavity. Idiopathic chylopericardium is extremely rare. It is characterized by chylous effusion in the pericardial cavity and clinical signs of cardiac tamponade. Conservative treatment of chylothorax is usually satisfactory, while treatment of chylopericardium needs to be surgical in the vast majority of patients. We present a unique case of idiopathic chylothorax and chylopericardium in a preterm neonate, successfully treated conservatively.*

Descriptors: CHYLOTHORAX; PERICARDIAL EFFUSION; INFANT, PREMATURE

## INTRODUCTION

Chylothorax may be congenital, or more often secondary to intrathoracic surgery, trauma or malignancy. Congenital chylothorax is usually idiopathic (1). However, it may be associated with lymphangiomatosis and lymphagiectasia, congenital hearth disease, Down's and Noonan's syndromes, tracheo-oesophageal fistula and polyhydramnios (1-3).

The characteristics of chyle are milky appearance, pH 7.4-7.8, specific gravity 1012-1025, lymphocytes 400 to 6800 cells/mm$^3$, and erythrocytes 50 to 600 cells/mm$^3$. Normal composition of chyle is: total protein 21-59 g/L, total fat 4-60 g/L, glucose 2.7-11.1 mmol/L, urea 1.4-3.0 mmol/L, and electrolytes similar to plasma values, while cholesterol/triglyceride ratio is < 1 (4).

Chylothorax may be bilateral, but more frequently it is unilateral, and on the right side (5). Treatment has to be considered in the light of the etiology and the patient's overall condition. Conservative treatment includes drainage of the pleural cavity, use of a low fat, high-protein diet, enriched with medium chain triglycerides (MCT) and total parenteral nutrition (TPN). Somatostatin, or its analogue octreotide has been useful in the treatment (6). Surgical intervention is indicated when there is no resolution of chyle flow after up to 4 weeks of conservative treatment (6).

Chylopericardium, chylous effusion in the pericardial cavity is an even more rare condition. It can occur after surgery of congenital heart disease, as a consequence of thoracic surgery or because of chest trauma or mediastinal neoplasms (7). Idiopathic chylopericardium is extremely rare, especially in neonates. In this report we present a unique case of congenital idiopathic chylothorax followed by idiopathic chylopericardium in a preterm neonate.

## CASE REPORT

A 1260 g male infant was born at 29 weeks gestation. Six days prior to delivery the mother was hospitalized because of vaginal hemorrhage. On delivery the infant was vigorous, and his Apgar score was 9. Ten hours after birth the infant had episodes of apnea, and was transferred to the Pediatric Intensive Care Unit (PICU). On admission his general condition was good. He was fed through a gastric tube. An umbilical venous catheter was inserted. However, three days after admission to the PICU he became dyspnoic and cyanotic. A chest radiograph revealed a left-sided liquidothorax. He was intubated and mechanical ventilation was started. After initial thoracocentesis, an intercostal drain was inserted. Biochemical and cytological analysis of the milky drained fluid determined it to be chyle: total protein 31.1 g/L, glucose 5 mmol/L, cholesterol 0.2 mmol/L, triglycerides 13.8 mmol/L, L 3,000 cells/mm$^3$, neutrophile 2%, limfocyte 91%. Echocardiography revealed an atrial septal defect. The karyotype was normal and serological findings (Toxoplasma gondii, Rubella, Cytomegalovirus, Herpes simplex, Epstein-Barr, Coxackie and Parvovirus B 19) were negative. The patient was put on a low-fat diet enriched with MCT, combined with parenteral nutrition. He responded well to this treatment and the accumulation of chyle decreased steadily. After 14 days there was no detectable chyle in the pleural space, his clinical condition was good, and the

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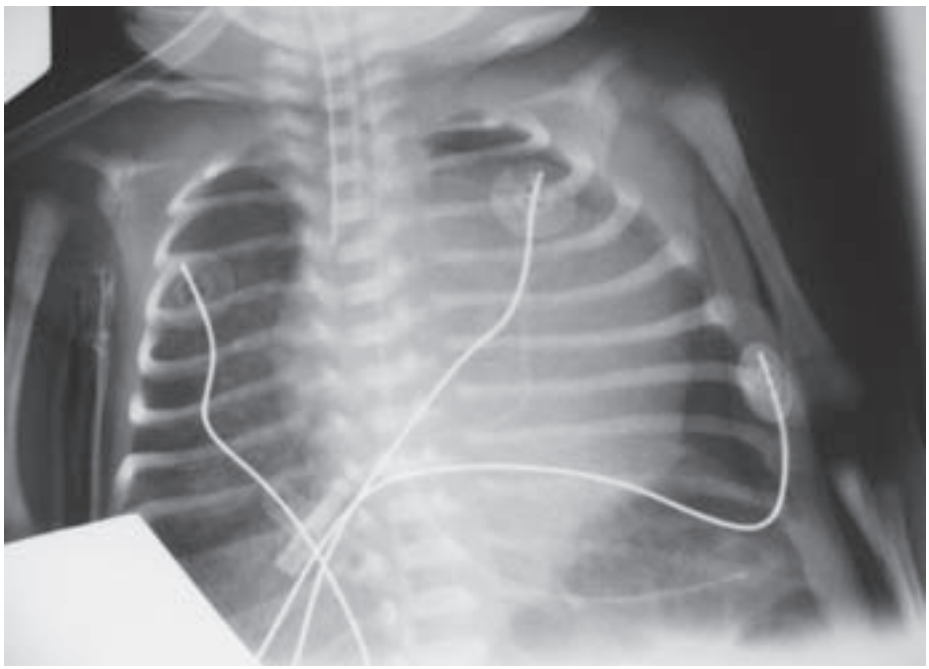

Figure 1. *Chest x-ray of a preterm neonate with chylopericardium*
Slika 1. *Rtg pluća nedonošćeta s hiloperikardom*

chest tube was removed. However, on the following day the clinical condition deteriorated again abruptly. Clinical signs caused suspicion of cardiac tamponade. The neonate was bradycardic with distant heart sounds. The diagnosis was proved by echocardiography and emergency pericardiocentesis was performed, 10 mL of fluid was removed, and a pericardial catheter was inserted. Chest radiography was also performed (figure 1). Biochemical and cytological analysis proved that the aspirated fluid was chyle: total protein 49 g/L, glucose 8.2 mmol/L, cholesterol 0.1 mmol/L, triglycerides 5.8 mmol/L, L 4,900 cells/mm$^3$; neutrophile 1%, limfocyte 90%. The CT scan after pericardiocentesis was unremarkable. Routine laboratory tests were within normal range, there were no signs of systemic inflammation, and cultures of the pericardial fluid were negative. Over the following 7 days, echocardiography was performed regularly. Total effusion volume was 70 mL. After 25 days of hospitalization the pericardial catheter was removed. Parenteral nutrition was gradually ceased, the patient was taken off mechanical ventilation and was breathing spontaneously. On day 42 he was transferred to the in-patient ward, and 6 days later he was discharged from hospital. Regular diet was instituted gradually and without deterioration. On the follow-ups, including 2 years after discharge from hospital, he was in a good condition and with normal growth parameters.

## DISCUSSION

Congenital idiopathic chylothorax is the most common cause of pleural effusion in the neonatal period (8). It is extremely important to recognize it early. Recognition and appropriate treatment are important to provide lung expansion, and to prevent hypoxic and chronic lung damage (5). Furthermore, severe nutritional depletion can result from continuous leakage of chyle (4). With the use of obstetric sonography, congenital chylothorax can be early recognized in the prenatal period. Elevated pleural fluid/serum IgG ratios may confirm diagnosis *in utero* (9). After birth, chest radiography is the first exam. The chylous characteristics of the fluid are confirmed by its lipid analysis: concentration of triglycerides is high, and cholesterol concentration is low. The treatment, recovery and long-term clinical and cognitive prognosis depend on the etiology and the patient's general condition (5).

In our patient, conservative treatment with drainage of effusion in the pleural space proved to be adequate. The conservative approach mandates careful evaluation of daily intravenous replacement of fluids, protein and electrolyte losses. The evolution of the disease was well controlled in our patient, until the occurrence of signs of cardiac tamponade. Whenever chylopericardium is diagnosed, it is extremely important to initiate adequate draining of the pericardial sac early on, in order to avoid serious consequences of tamponade. We considered chylopericardium in our patient as idiopathic, since CT revealed no abnormalities, and the response to conservative treatment was prompt and successful. Idiopathic chylopericardium has been noticed in both sexes, with most cases in children or young adults, and the majority of patients have to be treated surgically (10).

To our knowledge, this is the youngest ever reported patient with idiopathic chylopericardium, and the only case of idiopathic chylothorax followed with idiopathic chylopericardium in a preterm neonate ever reported. Conservative treatment of congenital chylothorax has a high success rate (8). However, the conservative treatment of idiopathic chylopericardium is rarely successful, and surgery is usually the treatment of choice (11). Our patient responded well to an initial conservative approach, and recovered completely with exclusive conservative treatment. Therefore, our experience may serve as an important starting instruction for management of neonates with idiopathic chylothorax and chylopericardium in the neonatal period (8, 11).

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KONZERVATIVNO LIJEČENJE IDIOPATSKOG HILOTORAKSA I HILOPERIKARDA U NEDONOŠČETA

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*Hilotoraks je rijetko stanje u kojem dolazi do nakupljanja hiloznog sadržaja u pleuralnu šupljinu. Idiopatski hiloperikard je iznimno rijedak. Kod njega dolazi do nakupljanja hiloznog sadržaja unutar perikarda s posljedičnom srčanom tamponadom. Konzervativno liječenje hilotoraksa obično je uspješno, za razliku od hiloperikarda koji u većine bolesnika zahtijeva kirurško liječenje. Prikazali smo jedinstven slučaj idiopatskog hilotoraksa i hiloperikarda u nedonoščeta koje smo uspješno konzervativno liječili.*

Deskriptori: HILOTORAKS; HILOPERIKARD; NEDONOŠČE

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